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Congenital pancreatic cyst: Preoperative diagnosis and management

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ABSTRACT

Background: Congenital pancreatic cyst is very rare in infants and children. The majority of pancreatic cysts in children are pseudocysts resulting from trauma, acute pancreatitis or infection.
Patients and methods: Three children with true congenital pancreatic cyst were treated. Their medical charts were reviewed for age at presentation, sex, clinical features, diagnosis and management.
Results: Three children (1 male and 2 females) with a mean age of 23 months (4 months-4 years) were treated for congenital pancreatic cyst. One of them presented with an abdominal mass while another presented with non-bile stained vomiting and abdominal distension. The third was diagnosed to have left sided Wilms tumor and during investigation was found to have congenital pancreatic cyst. CT-scan and MRI were helpful in confirming the diagnosis. All were treated surgically. One had total surgical excision while the other two had cystojejunostomy.
Conclusions: Congenital pancreatic cyst is very rare in the pediatric age group and should be considered in the differential diagnosis of cystic lesions of the pancreas in infants and children. Preoperative diagnosis is important to plan surgical intervention. Complete surgical excision is the treatment of choice but if this is not feasible a form of internal drainage, a cystoduodenostomy or a Roux-in-Y cystojejunostomy should be done.

1. Introduction

Congenital pancreatic cysts are extremely rare in infants and children and pose unique diagnostic and surgical challenges. Prenatal diagnosis is extremely rare and they are generally asymptomatic discovered as an incidental abdominal swelling. They can attain a large size and may cause abdominal distention or become symptomatic as a result of pressure on adjacent viscera [1–7]. Preoperative diagnosis is difficult especially when they reach a large size as they as they may be confused with other intra-abdominal cyst including pancreatic pseudocysts or duplication cyst of the pancreas [1,3,4]. The exact etiology of congenital pancreatic cyst is not known but it is accepted that they occur as a result of developmental anomalies of the pancreas and results from sequestration of primitive pancreatic ducts [6–9]. Once diagnosed, the treatment of congenital pancreatic cyst is surgical excision. This however is not always feasible and a variety of surgical procedures were described to treat congenital pancreatic cyst depending on its location. These include cystoduodenostomy, a Roux-en-Y cystojejunostomy, total cystectomy and total cystectomy with distal pancreatectomy for those located in the tail of the pancreas [1,2,4]. This report describes three cases of true congenital pancreatic cyst, outlining aspects of diagnosis and management. The literature on the subject is also reviewed.

2. Case reports

2.1. Case no. 1

A 4-month-old female was referred to our hospital because of non-bile stained vomiting and abdominal distension. The vomiting started immediately after birth and was on and off while the abdominal distension started two weeks prior to her presentation. It was small to start with and increased gradually. Clinically, she was found to have a large, smooth and non-tender abdominal

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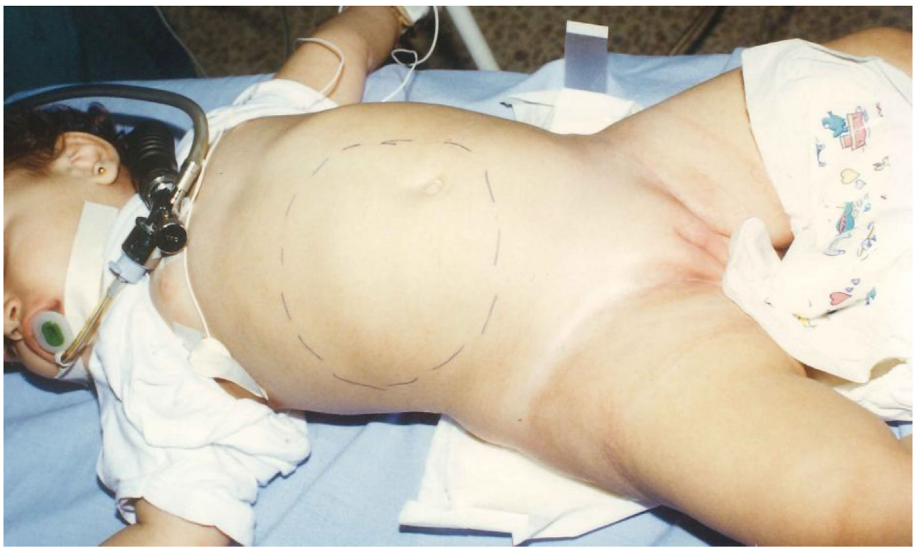

Fig. 1. A clinical photograph showing a female child with a large abdominal swelling.

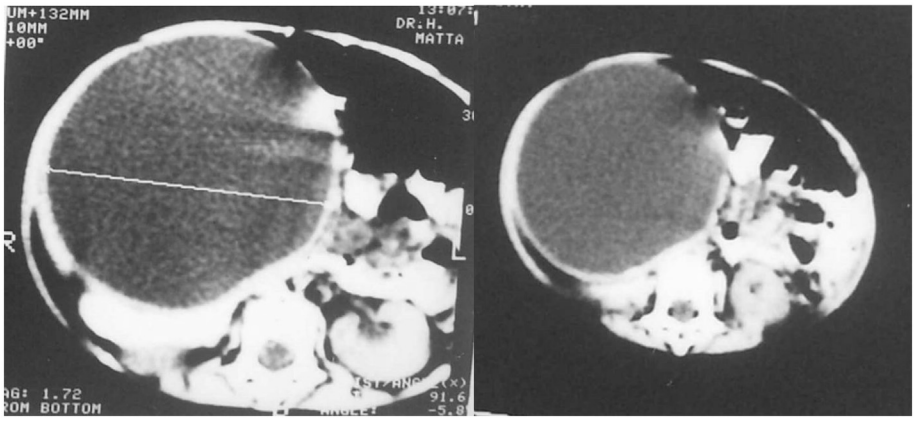

Fig. 2. A and B: Abdominal CT-scan showing a large right-sided abdominal cystic swelling.

swelling. The swelling occupied most of her upper abdomen (Fig. 1). She was admitted to the hospital and her investigations revealed a normal CBC, amylase, BUN and creatinine. Plain abdominal x-ray revealed a soft tissue density involving most of the upper part of the abdomen and pushing the bowels downwards. CT-scan of the abdomen showed a large cystic swelling involving most of the abdomen and measuring about 12 cm in diameter (Fig. 2A and B). The origin of the swelling could not be determined. She underwent exploration laparotomy and this revealed a large cystic swelling which was smooth to firm in consistency and pushing the intestines downwards and to the left side. The swelling was dissected and found to be arising from the head and body of the pancreas. Aspiration of the fluid in the cyst revealed an amylase level more than 5000 IU/L. The cyst could not be separated and removed from the pancreas and was drained via a Roux-in-Y cysto-jejunostomy. A biopsy taken from the cyst wall showed a cyst lined by cuboidal epithelium with mild inflammatory cell infiltrate. Postoperatively she did well, tolerated feeds and was discharged home two weeks postoperatively.

2.2. Case no. 2

A 1.5-year- old male child was referred to our hospital with

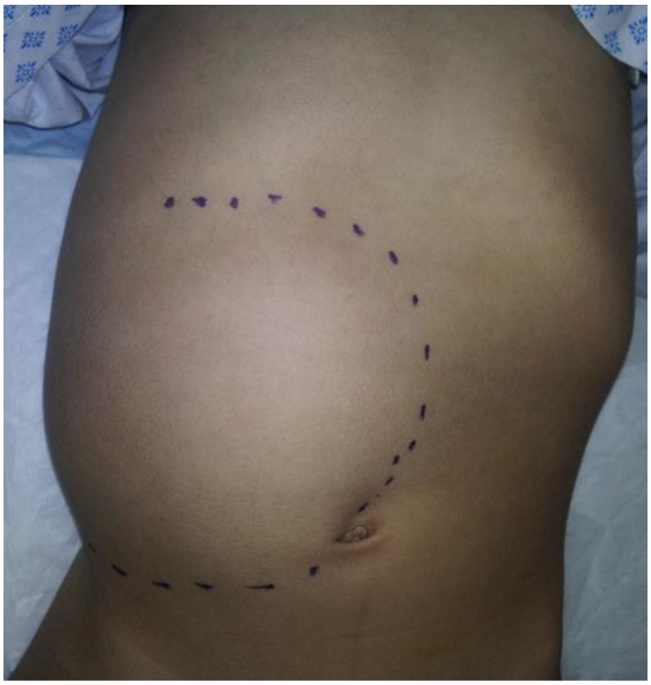

Fig. 3. A clinical photograph showing a large right-sided abdominal swelling.